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CASE REPORT

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# Thoracic segmental spinal anesthesia for radical nephrectomy in a patient with amyotrophic lateral sclerosis—a case report

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## Abstract

**Background** Amyotrophic lateral sclerosis (ALS) is a rare but fatal neurodegenerative disease of the motor neurons that causes muscle weakness, atrophy, and eventually respiratory failure and death. Anesthesia in these patients carries the risk of aspiration, altered response to muscle relaxants and opioids, ventilatory depression, and neurotoxicity to local anesthetics. Thoracic segmental spinal anesthesia may be a viable alternative to general anesthesia in upper abdominal surgeries in ALS patients.

**Case presentation** A 38-year-old patient with ALS diagnosed with a left renal mass of mitotic origin was scheduled for radical nephrectomy after an evaluation. Thoracic segmental spinal anesthesia was planned for the patient. The surgery and perioperative period were uneventful, and postoperative analgesic consumption was minimal. The patient was discharged without any progress in his neurodegenerative state.

**Conclusions** Our experience with thoracic segmental spinal anesthesia for nephrectomy in this ALS has been satisfactory. Our goals of maintaining hemodynamic stability, avoiding respiratory depression, and limiting further neurological impairment were all achieved. Further studies are needed to establish thoracic segmental spinal anesthesia as a better modality for such cases.

**Keywords** Amyotrophic lateral sclerosis, Thoracic segmental spinal anesthesia, Nephrectomy, Respiratory depression, Neurodegenerative disease

## Background

Amyotrophic lateral sclerosis (ALS), or Charcot disease, is a rare progressive neurodegenerative disease of motor neurons. With neuronal degeneration, there is an interruption of signal transmission to the muscles, and the muscle gradually starts to weaken and undergo atrophy (Wijesekera and Nigel Leigh 2009). The onset of ALS is so subtle that it may be overlooked initially. The first sign usually appears in the limbs, manifesting as difficulty in doing simple tasks such as writing and walking. This is referred to as limb onset ALS. When an individual notices speech or swallowing difficulty, it is termed "bulbar onset ALS." With time, muscle weakness and atrophy progress to the rest of the body, manifesting as problems with mobility, swallowing, speech production, and breathing. Patients are usually depressed and anxious, and a form of dementia may develop over time (Zarei et al. 2015). The disease progresses and eventually causes death due to respiratory failure. Administration of anesthesia in such cases is still a major challenge and requires a multidimensional approach. Herein, we describe our

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experience with thoracic segmental spinal anesthesia in a patient with ALS posted for radical nephrectomy.

## Case presentation

A written and informed consent was obtained from the patient for the publication of this case report.

A 38-year-old male (weight of 63 kg and height of 168 cm) came to our hospital with complaints of severe pain in the left flank. He was diagnosed with ALS 3 years ago. His symptoms started as weakness in the right leg, which progressed steadily to all other limbs. After 2 years of increasing weakness and spasticity, he became completely bedridden. He also developed bulbar symptoms like severe dysarthria, dysphonia, and dysphagia over the last 9 months. A neurological examination revealed severe spasticity in all limbs along with widespread wasting of all large and small muscles. Tongue atrophy and fasciculation were present. Uvular deviation along with palatal arch asymmetry was noted. The sensory system was intact. Tendon reflexes in all limbs were brisk, along with a positive Babinski sign. He had motor power of 3/5 in all limbs. There was no bladder or bowel involvement. His single breath count was less than 15 s. He had a reduced mouth opening, modified Mallampati class 3, and limited head extension. His MRI brain and cervical spine showed no abnormalities. EMG showed fasciculations of the left abductor pollicis brevis, left tibialis anterior, paraspinal muscles (thoracic), and tongue suggestive of neurogenic involvement of skeletal muscles.

His routine blood investigations along with chest X-ray, ECG, and echocardiogram were normal. Pulmonary function testing was not feasible because of his bulbar symptoms. The CECT abdomen showed a left renal mass (44×55 mm) with enlarged perinephric lymph nodes (Fig. 1A).

Radical nephrectomy was planned, and informed consent was obtained after the risks associated with this type of anesthesia and surgery were explained. After a fasting period of 8 h, the patient was taken up for surgery. His preoperative vitals were BP 120/76 mmHg, pulse rate 70/min, respiratory rate 16–18/min, and $SpO_2$ 98% at room air. Thoracic segmental spinal anesthesia was planned after discussion with the surgical team, with general anesthesia as the fallback plan. Standard monitoring was instituted. Under aseptic conditions, a paramedian approach with a 23-G spinal needle in an assisted sitting position (Fig. 1B) was used to perform spinal anesthesia at the T7–T8 vertebral interspace, and 2 ml of 0.5% bupivacaine heavy with 120 µg of buprenorphine was injected. The patient was made supine with a 20° head-down tilt. His sensory and motor blocks were assessed by pin-prick and the modified Bromage scale (Kim et al. 2014). After 15 min, the patient attained a sensory block level of T6–L2 and Bromage score of 2. The patient was turned to the left lateral position with a wedge behind the flank. Supplemental oxygen was given with a face mask at 4 l/min. Injections of cefuroxime 1.5 gm, tranexamic acid 500 mg, and midazolam 1 mg were administered intravenously. Blood loss was 300 mL, and urine output was 700 ml. One unit of PRBC and 1000 ml of RL were transfused. The surgery lasted 2 h. The patient's postoperative sensory level regressed to T8–T12 level when he was assessed before shifting to the post-anesthesia care unit. Complete block regression was seen 4 h after surgery.

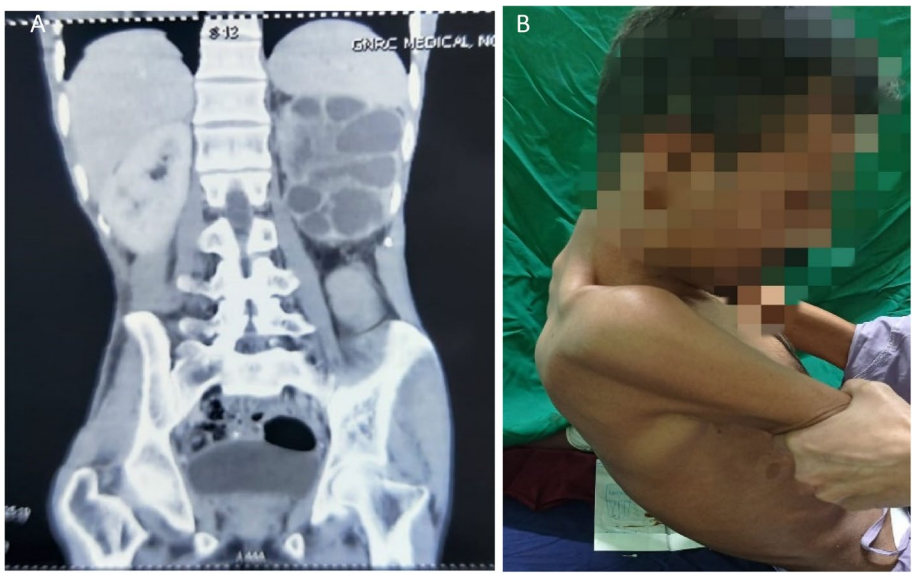


**Fig. 1** **A** Left renal mass seen in the CECT abdomen scan. **B** Patient with severe spasticity in assisted sitting position

Postoperative pain was managed with the infusion of paracetamol. The patient was shifted from the hospital at the end of the 5th day. No neurological complications were noted during the hospital stay or till one month of follow-up.

## Discussion

Anesthesia for surgery in patients with ALS has always been challenging. Risk of aspiration, increased sensitivity and uncertain response to muscle relaxants and opioids, ventilatory depression with postoperative respiratory failure, and neurotoxicity to local anesthetic agents are major concerns. Our goal was to minimize hemodynamic fluctuations, avoid airway manipulation, limit opioid consumption, and avoid using neuromuscular blockers. After discussing the procedural risks and benefits with the patient party and the surgical team, thoracic segmental spinal anesthesia was planned for this case. Thoracic epidural anesthesia was avoided due to technical difficulty and the requirement of intense skeletal muscle relaxation in this patient.

General anesthesia in ALS patients has a high risk of gastric fluid aspiration. Opioids, sedatives, and neuromuscular blockers in these patients carry a possible risk of increased sensitivity that may cause respiratory depression, delayed reversal, and prolonged postoperative ventilation (Rosenbaum et al. 1971). The complications of general anesthesia in ALS were highlighted by Hoeper et al. In their study cohort of 78 patients, 3 cases of prolonged ventilation, 1 case of post-extubation respiratory distress requiring tracheostomy, and 4 cases of prolonged hospital stay were noted (Hoeper et al. 2019).

Uneventful spinal-epidural anesthesia with non-invasive ventilation has been reported by Kock-Cordeiro et al. in a case of ALS undergoing cesarean section (Kock-Cordeiro et al. 2018). Epidural anesthesia may cause less toxicity to the nerve due to a lower concentration of drugs directly acting on the dorsal horn cells of the spinal cord (Hebl et al. 2016). But its administration requires technical expertise and proper positioning, which is difficult in ALS because of severe spasticity.

Thoracic segmental spinal anesthesia is a viable alternative to general anesthesia and has distinct advantages over conventional spinal anesthesia. The amount of cerebrospinal fluid is less, and the nerve rootlets are smaller in the thoracic segment of the spinal cord. So, an excellent surgical field pertinent to the dermatome level with an intense sensory block with less motor blockade can be achieved with a lower amount of local anesthetic and opioids as the drug dilution per segment from the site of injection is reduced (Hogan et al. 1996). Subclinical autoimmune dysfunction and blockade of cardio-acceleratory fibers after regional anesthesia may produce profound hypotension and bradycardia in these patients, causing cardiovascular collapse (Shemisa et al. 2014). Proper preloading and careful positioning reduce the major hemodynamic fluctuations. Extensive sympatholysis is not achieved in thoracic segmental spinal anesthesia unlike in conventional spinal anesthesia and adds to further hemodynamic stability. Early mobilization achieved here adds to patient satisfaction and healthy response in the postoperative period (Zundert et al. 2006).

Worsening of neurological symptoms in ALS patients has been reported after spinal anesthesia (Dripps and Vandam 1956). The lack of a protective nerve sheath and spinal cord-associated demyelination make the spinal cord more susceptible to the neurotoxic effects of local anesthetics (Guay 2008). Iatrogenic complications from epidural anesthesia like needle trauma, nerve ischemia, and epidural hematoma can further compromise the neurological state (Kopp et al. 2015). On the other hand, Hebl et al. found no evidence of neurological deterioration in their study on the effects of spinal anesthesia in a group of 139 patients with previous CNS disorders, which included 5 cases of ALS (Hebl et al. 2016). Panchamia et al. reported an ALS case undergoing right total hip arthroplasty under spinal anesthesia without any neurological complications (Panchamia et al. 2020). The postoperative neurological deficit in ALS patients reported earlier had a complex etiology. Recently, surgical stress-induced inflammatory neuropathy has also been cited as a possible cause (Ahn et al. 2011). No early or delayed neurological deterioration was observed in our case till 1 month of follow-up.

## Conclusions

Thoracic segmental spinal anesthesia demands careful preoperative assessment by an experienced anesthetist. Our experience with this technique for various surgeries in patients with pulmonary comorbidities helped us to manage this case uneventfully. We feel this can be a safe alternative anesthetic modality in patients with ALS. By reporting this case, we aim to highlight the need for a wider evaluation and further study.

### Abbreviations

| | |
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| ALS | Amyotrophic lateral sclerosis |
| ECG | Electrocardiogram |
| CECT | Contrast-enhanced computed tomography |
| MRI | Magnetic resonance imaging |
| EMG | Electromyography |
| CNS | Central nervous system |
| RL | Ringer's lactate |
| PRBC | Packed red blood cells |

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**Authors' contributions**
PH: conduct of the case and patient follow-up, literature search, and manuscript preparation. RB: conduct of the case, literature search, and manuscript preparation. AB: conduct of the case and manuscript review. All authors have read and approved the final manuscript.

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**Availability of data and materials**
The datasets used along with the supplementary images are available with the corresponding author.

## Declarations

**Ethics approval and consent to participate**
Not applicable.

**Consent for publication**
Written informed consent from the patient was obtained for the publication of the case report including case details and images in the manuscript.

**Competing interests**
The authors declare that they have no competing interests.

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